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Acute diffuse peritonitis due to spontaneous rupture of a primary gastrointestinal stromal tumor of the jejunum: A case report

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ABSTRACT

INTRODUCTION: Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract. Overt peritonitis caused by GIST rupture is very uncommon. Three types of GIST rupture have been described: closed perforation due to abscess (abscess type), hemoperitoneum leading to rupture of the hematoma capsule in the tumor (hemoperitoneum type), and perforation of the digestive tract via a fistula leading to central necrosis of the tumor (bowel perforation type). This report describes a patient with spontaneous tumor rupture and diffuse peritonitis, a variant of the bowel perforation type of GIST rupture.

PRESENTATION OF CASE: A 74-year-old man presented with symptoms of vomiting and abdominal pain. Computed tomography (CT) scan revealed an approximately 10 × 7-cm mass in the pelvis with free air and fluid collection. Emergency laparotomy revealed a tumor in the jejunum, which was ruptured with a hole measuring 5 mm in diameter. The tumor and part of the jejunum were resected. Immunohistochemically, the mass was diagnosed as a GIST originating from the gastrointestinal tract. Despite chemotherapy with imatinib mesylate, the patient died 22 months after surgery.

CONCLUSIONS: This report describes a patient with acute diffuse peritonitis due to spontaneous rupture of a primary GIST of the jejunum.

1. Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumor of the gastrointestinal tract [1]. The clinical symptoms of GIST complications include vague abdominal pain, hematemesis, and intestinal obstruction. However, overt peritonitis caused by GIST rupture is very uncommon [2]. Three types of GIST rupture have been described to date: closed perforation due to abscess (abscess type), hemoperitoneum leading to rupture of the hematoma capsule in the tumor (hemoperitoneum type), and perforation of the digestive tract via a fistula leading to central necrosis of the tumor (bowel perforation type). This report describes a patient with spontaneous tumor rupture and diffuse peritonitis, a variant of the bowel perforation type of GIST rupture. This case report has been prepared in line with the SCARE criteria [3].

2. Presentation of case

A 74-year-old man presented with symptoms of vomiting and abdominal pain for 10 h. His body temperature was 38.1 °C; blood pressure, 144/91 mmHg; radial pulse rate, 118 beats/minute. Abdominal examination revealed tenderness and muscular defense in the epigastric fossa. Blood tests showed a white blood cell count of 7470/mm^3, and a C-reactive protein concentration of 6.39 mg/dL. Computed tomography (CT) scan revealed a solid mass measuring approximately 14 cm × 7 cm within the pelvis, and free air around the gastric cardia and perihepatic fluid (Fig. 1a–c). These findings suggested peritonitis induced by organ rupture. Emergency laparotomy revealed an approximately 14-cm solid tumor in the jejunum, located 100 cm from Treitz's ligament (Fig. 2). The tumor was perforated, with a hole measuring approximately 5 mm. Segmental

Abbreviations: CA 19-9, cancer antigen 19-9; CEA, carcinoembryonic antigen; CT, computed tomography; MRI, magnetic resonance imaging; PET, positron emission tomography; GIST, gastrointestinal stromal tumor; H&E, hematoxylin and eosin.

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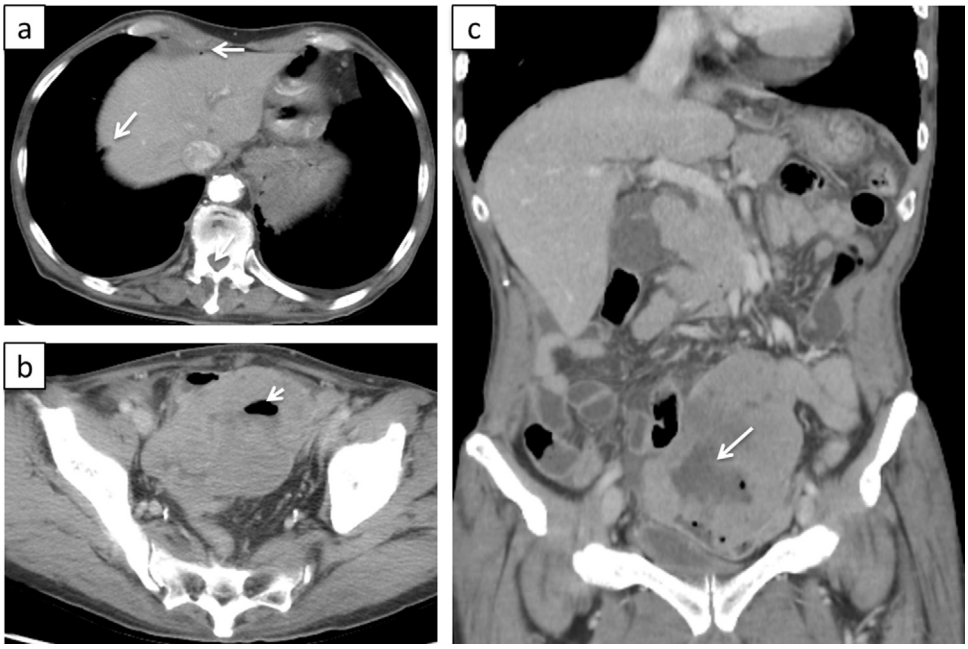

Fig. 1. Computed tomographic scans, showing (a) free air at the liver surface and the gastric cardia with fluid collection (white arrows) and (b, c) a solid mass within the pelvis, measuring approximately 14 cm × 7 cm, with some air (short arrows) and internal liquid (long arrows).

resection of the jejunum with the tumor showed that the resected mass was a well-circumscribed tumor, measuring 14 cm × 7 cm and penetrating the jejunum. The solid parenchyma contained an area central necrosis with a fistula to the lumen of the jejunum. (Fig. 3) Hematoxylin and eosin staining showed proliferation of spindle-shaped cells, and immunohistochemical staining showed that the tumor was positive for c-kit and CD34, with approximately 10% of the tumor cells positive for nuclear expression of the proliferation-associated antigen Ki-67. (Fig. 4) The patient was diagnosed with a high-risk GIST of the jejunum. Because of cardiac insufficiency and pneumonia, the patient could not be treated with adjuvant imatinib. A follow-up CT 1 year after surgery showed the appearance of liver metastases. Chemotherapy was initiated with a lower dose of imatinib mesylate, but the treatment was discontinued 3 months later due to cough and general malaise. The patient died 22 months after surgery.

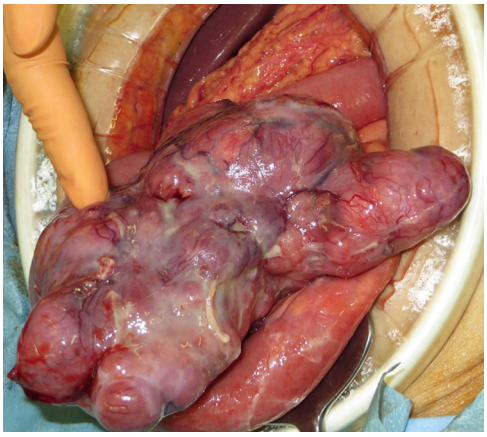

Fig. 2. Intraoperative photograph, showing an approximately 14 cm perforated tumor with a hole measuring approximately 5 mm (white arrow) in the jejunum. The tumor was located 100 cm from Treitz's ligament.

3. Discussion

GISTs are visceral tumors arising from any site within the gastrointestinal tract. Approximately 60–70% of these tumors occur in the stomach, 25–35% in the small intestine, and 10% in the jejunum, whereas the esophagus, colon, rectum, and appendix are rarely affected [4]. GISTs are usually associated with abdominal pain, palpable mass, and/or GI bleeding, accompanied by fever, anorexia, weight loss, and/or anemia [5]. Approximately 10% of GISTs are located in the jejunum [6]. The most commonly reported clinical symptoms are bleeding and obstruction [7]. However, GISTs originating from the small bowel rarely cause perforation or cause acute diffuse peritonitis. Common symptoms of small intestinal GISTs include abdominal pain and a palpable mass, with early satiety and abdominal fullness also occurring frequently [8]. Fever, anorexia and weight loss are rarely observed [9]. A MEDLINE search, using the key-words "GIST" AND "small intestine" AND "perforation" or "rupture," revealed only 15 cases of small intestinal GIST with perforation; including the present case (Table 1). Three types of GIST rupture have been identified: abscess; hemoperitoneum; and bowel perforation types.

Abdominal pain is a chief symptom in patients with GIST. Statistical analysis revealed that 58% of patients with gastric GISTs experienced bleeding in the digestive tract and 61% experienced abdominal pain; these rates are higher than those in patients with GISTs at other locations are [10]. In contrast, acute abdominal symptoms were more frequent in patients with jejunal (40%) and ileal (60%) GISTs than in those with gastric GISTs ($p < 0.05$ each) were [10]. Patients with the hemoperitoneum type of GIST rupture frequently experience hypovolemic shock, short loss of consciousness, and sudden abdominal pain, whereas patients with the bowel perforation type of GIST rupture experience diffuse, worsening abdominal pain. Symptom duration is longer in patients with the abscess type than in those with the other types of GIST rupture (Figs. 3 and 4).

To date, all tumors in patients with GIST rupture originated in the jejunum, ranging in size from 6.5 cm to 21 cm. No differences in tumor origin and size have been observed among patients with the three types of GIST rupture. GIST perforations are more